An unusual cause of death in Wegener's granulomatosis

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Mortality among patients with Wegener's granulomatosis (WG) is more likely among those with renal disease.[1 2] Among the non-renal causes, laryngeal involvement with airway obstruction is a rare but important preventable cause of death.[3] Here, we describe a patient with WG and laryngeal involvement, in whom death occurred by inhalation of a nasal crust.

The patient, a 26 year old woman, first presented to us in 1998 with a 3 year history of intermittent stridor associated with hoarseness of voice, intermittent nasal stuffiness, nasal crusting, and recurrent sinusitis. Serum perinuclear antineutrophil cytoplasmic antibody was positive at a titre of 1/80 by indirect immunofluorescence, with myeloperoxidase-ANCA of 28 AU/ml (normal range 0–5) by enzyme linked immunosorbent assay (ELISA), while cytoplasmic ANCA was negative and proteinase 3-ANCA was normal. Examination with fibre optic nasendoscopy demonstrated abnormal tissue in the subglottic region (in retrospect, thought to be granulomatous tissue). A biopsy specimen from her nasal mucosa showed necrotising vasculitis in keeping with a diagnosis of WG.

Chest radiography and a high resolution computed tomographic scan of her chest showed nodules in both lung fields. There was no evidence of renal or other systemic disease.

She was treated with prednisolone and methotrexate, to which she responded well (Birmingham Vasculitis Activity Score (BVAS)[4] improved from 11 to 0 (range 0–63)). However, she reported that the nasal crusts were ''falling down to the back of her throat'' and were causing her occasional difficulty in breathing. She also had recurrent nasal infections with *Staphylococcus aureus* requiring frequent courses of antibiotics.

In December 1999, she developed sudden onset of breathlessness associated with generalised seizures and loss of consciousness. She recovered rapidly after a brief period of ventilation by the ambulance crew. During intubation it was noted that she had inhaled a nasal crust, which had blocked off an already narrowed trachea. Her WG was in remission at this stage (BVAS 0). A computed tomographic scan of her subglottic region showed soft tissue mucosal thickening affecting the subglottic region, the appearance of which raised the possibility of cartilage replacement by granulomatous disease. She was referred to the otolaryngologist for consideration of local subglottic steroid injection, but this was not required as her stridor resolved within a few days, and direct laryngoscopy performed a few months later did not show any evidence of subglottic or tracheal stenosis.

She remained well until April 2004, when she re-presented with sudden onset of breathlessness and loss of consciousness. The ambulance crew initiated cardiopulmonary resuscitation for asystolic cardiac arrest. They were unable to pass a cuffed endotracheal tube owing to a mass occluding the narrowed airway. On arrival in the hospital, she had a Glasgow Coma Scale of 3 with no respiratory effort. She was intubated with an uncuffed endotracheal tube and placed on continuous positive airway pressure ventilation but died 48 hours later. Although a postmortem examination was not performed, the likely cause of her death was occlusion of her narrowed trachea caused by inhalation of a nasal crust.

This is the first reported case of mortality in WG due to inhalation of a nasal crust. The development of subglottic stenosis is a potentially life threatening complication and hence, several reports have emphasised the importance of meticulous care of the upper airway in such patients.[5–7] Our case highlights the fact that care of the upper airway in patients with subglottic stenosis should not only include paying attention to the subglottic region but also to the nose and paranasal sinuses, as complete obstruction of the already narrowed airway can occur from impaction of nasal crusts. In view of this presentation, we recommend more careful attention to extensive nasal crusting in patients with subglottic stenosis, through increased input from otolaryngologists. It is, however, difficult to know if this would prevent further similar fatalities in future.

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